Neonatal Asphyxia and Functional Tricuspid Atresia*

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The patient was a 2,700 gm full term female, the product of a normal 41-week pregnancy and delivery. She was noted to have cyanosis and labored breathing at birth. The Apgar scores were 5 and 6 at 1 and 5 minutes respectively. The heart sounds were heard best on the right side of the chest. No murmur was present. The breath sounds were poor and bowel sounds were audible in the left chest. The abdomen was scaphoid and the liver was not palpable. There was hypoplasia of the left arm and hand with a contracture of the left axilla. Chest roentgenogram revealed the presence of the liver and bowel loops in the left chest. The baby was intubated and placed on a respirator in 100 percent O₂. The pH was 6.6, Po₂ was 20, and the Pco₂ was 85 mm Hg. The baby was treated with trishydroxyaminomethane (THAM). At operation, a left-sided foramen of Bochdalek hernia and hypoplasia of the left lung were found. The hernia was reduced and the diaphragmatic defect repaired. The patient remained severely hypoxic and acidic despite maximal ventilatory pressures, 100 percent oxygen, and therapy with THAM. In an attempt to decrease the pulmonary vascular resistance, the patient was brought to the cardiac catheterization laboratory for infusion of tolazoline into the pulmonary artery. A French No.

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4 Berman angiographic catheter was advanced to the right atrium, left atrium, and left ventricle, but could not be inserted into the right ventricle despite numerous attempts. AP angiography was therefore performed with injection into the right atrium (Fig 1). The infant expired two hours after catheterization.
Diagnosis: Functional tricuspid atresia secondary to severely elevated pulmonary vascular resistance in a newborn.

Figure 1 shows that the contrast material injected into the right atrium had passed to the left atrium and left ventricle. No contrast material was visible in the right ventricle (RV) which appeared as a triangular filling defect, bounded by the contrast-filled right atrium (RA), left atrium (LA), and left ventricle (LV). This angiographic picture has been described as diagnostic of tricuspid atresia.\(^1\) Identical pressures in both atria with right-to-left shunting across the foramen ovale further supported this diagnosis. At autopsy, however, both ventricles, atrioventricular valves, and semilunar valves were normal. There was dilatation of the right ventricle and tricuspid valve.

Infants with neonatal asphyxia, particularly in association with a diaphragmatic hernia, often have increased pulmonary vascular resistance. This may result in right-to-left shunting at the level of the foramen ovale or ductus arteriosus.\(^2\) If the resistance is severe, forward flow across the pulmonary valve may be minimal. Freedom et al\(^3\) described a group of patients in whom no forward flow was seen across the normal pulmonary valve on right ventricular angiography because of high pulmonary arteriolar resistance.

In our patient, it is most likely that extremely high pulmonary vascular resistance caused by perinatal asphyxia prevented forward flow from the right ventricle, so that all or most of the opacified asphyxia entered the left atrium, and no filling of the right ventricle took place. This produced the misleading angiographic picture of tricuspid atresia. Differentiation between structural and functional tricuspid atresia may be difficult.

References
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